heart. J Cardiovasc Surg 1992;33:570.
5. Ceresoli GL, Ferreri AJ, Bucci E, Ripa C, Ponzoni M, Villa E. Primary cardiac lymphoma in immunocompetent patients. Cancer 1997;80:1497-506. http://doi.org/cd99hg
6. Petrich A, Cho SI, Billett H. Primary cardiac lymphoma. An analysis of presentations, treatment and outcome patterns. Cancer 2011;117:581-9. http://doi.org/d3sxbr

REV ARGENT CARDIOL 2016;84:249-250. http://dx.doi.org/10.7775/rac.v84.i3.8056

Secondary Cardiac Plasmacytoma

Malignant cardiac tumors are rare and, among them, cardiac plasmacytoma is extremely uncommon.

We present the case of a 68-year-old woman with multiple myeloma (MM) under chemotherapy treatment, who was admitted to the emergency department due to a 3-month history of episodes of dyspnea and symptoms suggestive of a heart condition. A PET-CT scan had previously revealed multiple hypermetabolic masses mainly in the right chambers, consistent with an active cancer process. A magnetic resonance imaging (MRI) was performed for better anatomical mapping of the interference of these masses with cardiac motility.

In addition, the purpose of this work is to present a brief update of this uncommon heart condition, and to highlight the usefulness of imaging methods, especially PET-CT scan and MRI, which have been shown to be valuable for proper diagnosis and follow-up.

The patient consulted for a 3-month history of dyspnea. A control PET-CT scan revealed multiple hypermetabolic lesions located mainly in the heart and affecting the right adrenal gland, the right pelvic cavity, and the abdominal wall (with no evidence of bone hypermetabolic lesions), consistent with an active cancer process (Figure 1). In view of this condition, MRI was suggested for better anatomical discrimination, and to study the cardiac dynamics affected by these masses.

The study was performed using a Siemens Avanto 1.5T system (Siemens, Erlangen, Germany) with surface coils and ECG gating. All the images were obtained during a simple apnea. Anatomical spin-echo, STIR sequences and functional SSFP (True FISP) cine sequences were performed in 2, 4 and 3 chamber cardiac axes, and short-axis sequence from base to apex.

Image analysis showed multiple isointense, pedunculated masses with respect to the myocardium in SSFP cine-MRI and T1 sequences, behaving as hyperintense in STIR sequences.

Two pedunculated masses were identified in the interventricular septum, one of them prolapsing towards the left ventricular outflow tract during systole. Two images in the right atrium were also observed, one of them implanted in the tricuspid annulus and prolapsing towards the ventricular chamber.

STIR images (Figure 2) showed hyperintense focal areas in mid inferior, mid inferolateral, and basal anteroseptal segments, which would be associated to myocardial infiltration areas due to the tumoral process. Despite these findings, biventricular systolic function was preserved.

Diagnosis was made through leukemia and lymphoma immunophenotyping of the cardiac masses by flow cytometry, revealing a cytometric profile consistent with moderate size LNH B CD10+. These findings were consistent with plasma cells and suggestive of plasmacytoma.

Multiple myeloma (MM) is characterized by uncontrolled proliferation of plasma cells, generally restricted to the bone marrow. (1) Extramedullary MM spread occurs when malignant plasma cells form tumors (plasmacytomas) in other parts of the body outside the bone marrow, whose reported incidence in newly diagnosed MM ranges from 7% to 18% of patients, or 6% to 20% of cases during the course of the disease. (1-3)

The mechanisms of extramedullary spread in MM are poorly understood. Extramedullary spread in MM can have two different origins: direct extension from skeletal tumors when they disrupt the cortical bone; or hematogenous metastasis to distal organs. However, scientific evidence is not enough to demonstrate it. (1)

Extramedullary masses may arise as primary plasmacytomas in patients with normal bone marrow analysis, or manifest themselves in patients with confirmed diagnosis of MM (secondary plasmacytoma), the latter being more aggressive. (4-6)

According to published series, 80 to 85% of patients with extramedullary MM had plasmacytomas in muscles, tendons, fat, or digestive tract, and 15% of the cases in glands, liver, kidneys, respiratory airway, skin, or breast; cardiovascular system involvement was extremely rare or absent. (4, 5, 7) In a review of the literature, Keung et al. found 9 cases of extramedullary MM involving the heart followed by a report

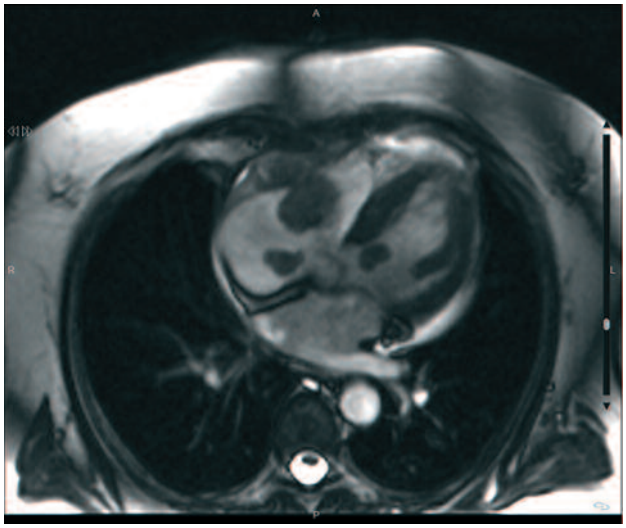

Fig. 1. See text for description.

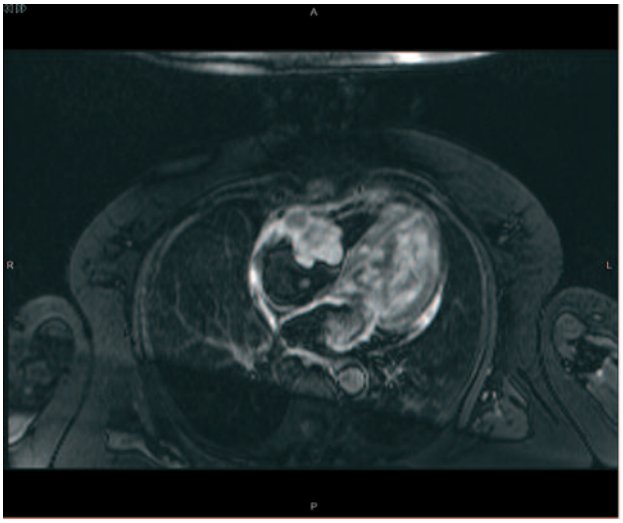

Fig. 2. See text for description.

of a case presenting as a cardiac emergency that required surgical intervention. (8)

The differential diagnosis should be made with all the cardiac masses involving the right heart, particularly metastases.

The extensive use of increasingly sensitive imaging techniques, such as CT scan, MRI, and PET-CT scan, have lately increased the diagnosed cases of extramedullary MM. (1, 7) MRI offers a full range of tools to localize, characterize and evaluate cardiac masses and their physiological effects. In addition, it allows studying the myocardial condition, establishing whether the myocardium has been infiltrated by the neoplasm in question. On the other hand, the PET-CT scan shows involvement at different levels, providing additional information that can be crucial for the treatment and follow-up of this condition. (6) The case we have reported demonstrates the usefulness of these techniques.

Laura C. Dragonetti[1], Leandro J. Carballo[1], Verónica Flores[2], Marcelo A. Pietrani[1], Ricardo D. García Mónaco[1]

[1] Department of Diagnostic Imaging, Hospital Italiano de Buenos Aires. Buenos Aires, Argentina

[2] Department of Diagnostic Imaging, Hospital San Francisco. Guayaquil, Ecuador

e-mail: laura.dragonetti@hospitalitaliano.org.ar

Phone: (0054 011) 4959-0200 - Fax: 4959-0200

REFERENCES

1. Bladé J, de Larrea CF, Rosiñol L. Extramedullary involvement in multiple myeloma. Haematologica 2012;97:1618-9. http://doi.org/bdv7

2. Bladé J, Fernández de Larrea C, Rosiñol L, Cibeira MT, Jimenes R, Powles R. Soft-tissue plasmacytomas in multiple myeloma: mechanisms of extramedullary spread, and treatment approach. J Clin Oncol 2011:29:3805-12. http://doi.org/bb589f

3. Varettoni M, Corso A, Pica G, Mangiacavalli S, Pascutto C, Lazzarino M, et al. Incidence, presenting features and outcome of extramedullary disease in multiple myeloma: a longitudinal study on 1003 consecutive patients. Ann Oncol 2010;21:325-30. http://doi.org/fp3vtc

4. Andrea R, Irene C, Armando F, De Vivo D, Simonetti G. Primary extramedullary plasmacytoma of the heart: a rare manifestation of plasmacellular tumor. Case Rep Radiol 2013;2013:290849. http://doi.org/bdv6

5. Chim CS, Loong F, Ma ES, Cheung W, Chan RH, Ooi GC. Plasma cell problems: Case 2. Extramedullary cardiac plasmacytoma presenting with cardiac tamponade. J Clin Oncol 2005;23:3140-3. http://doi.org/cqt2bj

6. Vrettou AR, Heffner LT, Rossi PJ, Clements SD Jr. Cardiac plasmacytoma: a rare clinical entity. Tex Heart Inst J 2014;41:554-7. http://doi.org/bhhd

7. Varettoni M, Corso A, Pica G, Mangiacavalli S, Pascutto C, Lazzarino M. Incidence, presenting features and outcome of extramedullary disease in multiple myeloma: a longitudinal study on 1003 consecutive patients. Ann Oncol 2010;21:325-30. http://doi.org/fp3vtc

8. Champeaux AL, Blaser JL, Myers JB, Schachter DT. Multiple myeloma involving the myocardium and coronary vessels. Arch Pathol Lab Med 2000;124:910-2.

REV ARGENT CARDIOL 2016;84:250-251. http://dx.doi.org/10.7775/rac.v84.i3.8055